Parents' Guide to

Hemoglobin H Disease

California Department of Health Services
Newborn Screening Program
Genetic Disease Branch

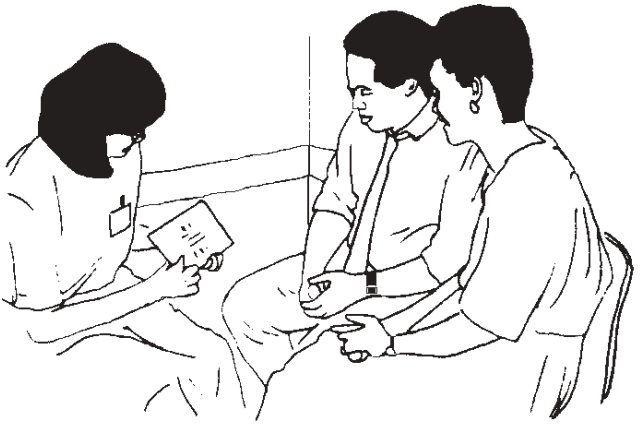
To Parents:

California State Law requires that all babies have the newborn screening test before leaving the hospital. A few drops of blood were taken from your baby's heel. One of the tests was for hemoglobin disorders.

Your health care provider may have recently told you that tests show that your baby has a blood disorder called hemoglobin H disease or hemoglobin H-Constant Spring disease.

This booklet was written to help parents learn more about hemoglobin H disease. People with hemoglobin H disease have varying symptoms.

Use this booklet to discuss this disorder with the specialists at a hematology center and to help you learn more about how to care for your child. A list of state approved hematology centers is included at the back in this booklet.



How Did My Baby Get Hemoglobin H Disease ?

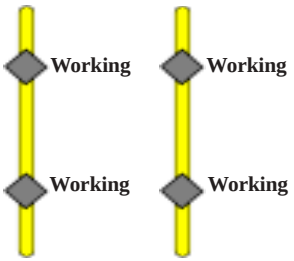
Hemoglobin H disease is a blood disorder that is inherited. Inherited means it is passed down from parent to child in the genes. Genes determine eye color, height, and other features including hemoglobin type. The baby received genes from both parents that resulted in the baby having hemoglobin H disease.

What is Hemoglobin?

Hemoglobin is found in the red blood cells. It gives the blood its red color and carries oxygen to all parts of the body. It is made up of iron and protein chains called globins. There are two kinds of globin in the usual adult hemoglobin called alpha globin and beta globin. The usual adult hemoglobin, called hemoglobin A, has 2 alpha chains and 2 beta chains.

What is Alpha Thalassemia?

Alpha thalassemia is a condition in which there is a decrease in the amount of alpha globin produced. Alpha globin is one of the protein chains that makes up hemoglobin. The amount of alpha globin produced is determined by the number of working genes. Most people have 4 working genes that make alpha globin.



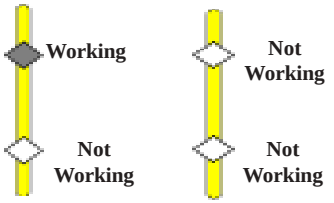
normal working genes

People with alpha thalassemia may have 0 to 3 working genes. The number of working genes determines the type of alpha thalassemia. People with 3 working genes are called ***silent carriers***. People with 2 working genes have ***alpha thalassemia trait***. People with only one working gene have ***hemoglobin H disease***. People with no working genes have ***alpha thalassemia major***. The more working genes missing, the more serious the type of alpha thalassemia.

Alpha thalassemia is more common in people from China, Southeast Asia (for example: Laos, Cambodia, and Vietnam), the Philippines, and other Asian countries, Mediterranean and Middle Eastern countries. However, people from any ethnic group can have a type of alpha thalassemia.

What is Hemoglobin H Disease?

People with hemoglobin H disease have only one working gene for alpha globin. They make less than the usual amount of alpha globin chains. Because of this the hemoglobin that they make is more unstable causing the red blood cells to break down more quickly. The result is fewer red blood cells, a condition called anemia.



one working gene - Hemoglobin H disease

Many people with hemoglobin H disease do not have serious health problems. However, since this disease affects a person's hemoglobin, they often have mild to moderate anemia which can cause them to be more tired. Sometimes the anemia can get worse and lead to other problems. However, these are rare except for children with hemoglobin H-Constant Spring disease which is a more severe form of this disorder.

What is Hemoglobin H-Constant Spring Disease?

Hemoglobin H-Constant Spring disease produces a longer than usual alpha globin chain. This causes the hemoglobin to be even more unstable than in hemoglobin H disease. This causes the red blood cells to break down faster than usual so there are less red blood cells in the body. This results in a more severe anemia. Other complications often include an enlarged spleen, gallstones, increased risk for infections, jaundice, and leg ulcers.

How Can I Care for My Baby?

The hematologist and other staff at the center will discuss how to care for your baby. Most people with hemoglobin H disease can lead relatively normal lives with proper treatment. Some may need occasional or ongoing blood transfusions. Children with hemoglobin H disorders are more likely than other children to get infections. Viral infections and fevers cause the red blood cells to break down faster, leading to anemia. To prevent problems call your baby's doctor or the hematology center whenever your child becomes ill.

Certain medications, household products, and fava beans must be avoided. They may cause severe anemia if your child comes in contact with them. These items may cause the red blood cells to break down faster. Treatment for people with hemoglobin H disorders may include taking a B vitamin called folic acid, avoiding the items listed on the next page and prompt treatment of infections. Iron overload can be a problem for the child getting blood transfusions. Your child should not be given iron medication unless the blood test shows he or she has iron deficiency anemia. Please check with your hematology center before giving iron supplements to your child.

What Should My Child Avoid?

On the next page is a list of medications and household products to avoid. Take this list with you to any doctor's appointment or emergency room visit. Tell the doctor your child has hemoglobin H disease. Show this list to any doctor seeing your child and to pharmacists who are filling prescriptions for your child. Do not give your child over-the-counter medicine without talking to your doctor about them first.

Should My Baby Avoid Anything Else?

Your child should avoid contact with fava beans and mothballs. Both of these can cause severe anemia. Do not have these in your home. Other types of beans such as red, black, and pinto beans or lentils are not a problem. Swallowing or inhaling mothballs can be very harmful to your child. Call your doctor immediately if your child swallows a mothball.

List of Medications and Products To Avoid*

Sulfa Drugs
Sulfacetamide (eye drops)
Sulfapyridine
Salfasalazine
(Salicylazosulfapyridine)
Sulfanilamide
Dapsone

Antimalarials
Primaquine
Chloroquine
Hydroxychloroquine sulfate

Other Antibacterials
Nalidixic acid (Negram)
Nitrofurantoins
Furazolidone
Chloramphenicol
Beta-amniosalicylic acid
Ciprofloxacin
Doxycycline

Analgesics
Aspirin
(Acetaminophen is safe as alternative)
Phenacetin
Acetanilide

Tuberculosis Drugs
Isoniazid
Rifampin

Folic Acid Antagonists
Pyrimethamine

Other Items
Iron Supplements [1]
Vitamin K analogues
Quinidine Gluconate
Phenazopyridine (pyridium)
Toluidine Blue (a dye)
Methylene Blue (a dye)
Naphthalene (Mothballs)

[1] *Unless laboratory proven iron deficiency.*

Source: Bull WHO 1989, Beutler 1994, updated by Michael Irvin, School of Pharmacy, UCSF, 1998

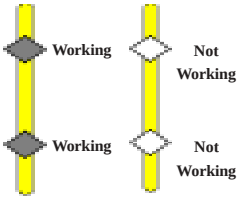
*The medical literature suggests that these medications and products may cause problems for people with hemoglobin H disease; however no studies have been conducted to prove that these substances are actually harmful for people with Hemoglobin H Disease.

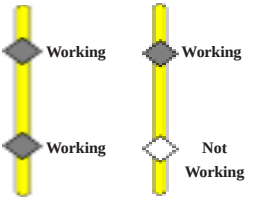
What Are the Chances of Having Another Child with Hemoglobin H Disease?

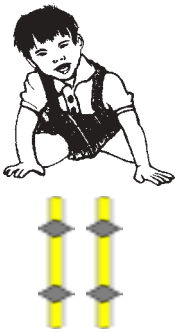
There are some combinations of alpha globin gene inheritance that can cause serious health problems. Special blood tests can tell you which combination you have and your chances of having another child with hemoglobin H disease in future pregnancies. Family testing should be discussed with the hematology center doctor.

For example, if one parent has alpha thalassemia trait and the other parent is a silent carrier, with each pregnancy, they have a 25% chance of having a child with 4 working alpha globin genes (***usual alpha globin***), a 25% chance of having a child with 3 working alpha globin genes (***silent carrier***),
a 25% chance of having a child with 2 working alpha globin genes (***alpha thalassemia trait***), and a 25% chance of having a child with one working alpha globin gene (***hemoglobin H disease***).

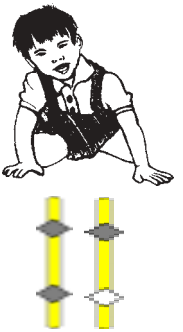
alpha thalassemia trait **silent carrier**



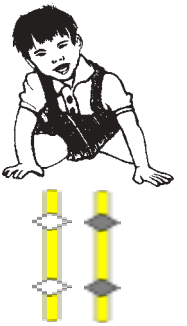




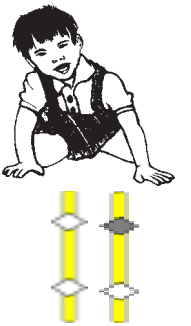
4 working genes (***usual alpha globin***)



3 working genes (***silent carrier***)



2 working genes (***alpha thalassemia trait***)



1 working gene (***hemoglobin H disease***)

What Is Your Chance of Having a Baby With Hemoglobin H Disease?

(Complete this with your genetic counselor after you have received your results.)

____ working genes

____ working genes

___ working genes

___ working genes

___ working genes

___ working genes

When Should I Call My Hematology Center?

A hematology center is a specialized center which has a team of experts who are trained in the treatment of hemoglobin disorders. They can provide your child with
the special care related to hemoglobin H disease.

If your baby experiences any of the following symptoms
of severe anemia contact your doctor or hematology center **immediately.**

- extreme fatigue
- pale and/or yellowish skin
- whites of the eyes become yellow
- stomach and/or back pains
- dark black stool
- dark orange urine

California Children's Services
Sickle Cell Diseases/Hemoglobinopathies Centers

Northern California

Alta Bates, Berkeley, 510/204-1609

UC Davis Medical Center, Sacramento, 916/734-2782

UC San Francisco General Hospital, San Francisco, 415/206-3770

Children's Hospital - Oakland, Oakland, 510/428-3372

Kaiser Permanente No. California, Oakland, 510/596-6592

Children's Hospital at Stanford, Palo Alto, 415/497-8953

Highland Hospital, Oakland, Oakland, 510/534-2055

Central California

Valley Children's Hospital, Madera, 559/353-5460

Saint Agnes Medical Center, Fresno, 559/449-5378

Southern California

City of Hope National Medical Center, Duarte, 626/359-8111 ext. 2915

Orthopedic Hospital, Los Angeles, 213/742-1402

Los Angeles County-USC Medical Center, Los Angeles, 323/226-7622

UC Los Angeles Medical Center, Los Angeles, 213/825-6708

Childrens Hospital of Los Angeles, Los Angeles, 323/669-4151

Cedars-Sinai Medical Center, Los Angeles, 310/855-4423

Kaiser Permanente Med. Ctr., So. California, Los Angeles, 323/857-4462

Harbor-UCLA Medical Center, Torrance, 310/222-4157

Long Beach Memorial Medical Center, Long Beach, 562/492-1062

Loma Linda Univ. Medical Center, Loma Linda, 909/799-5283

UC Irvine Medical Center, Orange, 714/456-8411

Children's Hospital of Orange County, Orange, 714/532-8636

UC San Diego Medical Center, San Diego, 619/543-5670

Notes And Questions

California
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Health Services

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